Coexisting Covert And Overt Menstruation

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Abstract

A 16 year-old unmarried girl presented with intermenstrual discharge per vagina. Examination and investigation revealed uterus didelphys, hemivagina and ipsilateral renal agenesis with haematocolpos. Patient was treated with drainage of the haematocolpos and excision of the vaginal septum.

INTRODUCTION

The reported incidence of Mullerian anomalies accounts for 0.5 - 5.0% in women¹. Uterine didelphys accounts for 11% of uterine malformations². Incidence of renal agenesis is 30% in Mullerian anomalies and more frequently with obstructed Mullerian anomalies². The specific association of uterus didelphys, obstructed hemivagina and an ipsilateral kidney was first described by Wilson in 1925³. This association is also known as Herlyn-Werner-Wunderlich Syndrome⁴. Recently it has been termed OHVIRA (Obstructive Hemivagina with Ipsilateral Renal Agenesis).

CASE REPORT

A 16 year-old, unmarried girl presented vaginal discharge after menstruation and cyclical lower abdominal pain for 2 months. There was a history of similar complaints 1 year back for which the patient consulted a local doctor and was diagnosed with pyometra. She attended menarche at 12 years-old and had regular menstrual cycles. She did not have significant past or family history. On examination patient had normal secondary sexual characteristics. Per vaginal examination revealed tender, tense & cystic bulge in the right anterolateral vaginal wall ~ 7 x 8 cm size. Our clinical diagnosis was vaginal wall cyst or a Hematoma. Transabdominal sonography revealed double uterus (Fig.1) with obstructed hemivagina and ipsilateral renal agenesis. The findings were confirmed with MRI (Fig.2 & Fig.3)

Figure 1 Figure 1





Figure 2 Figure 2



Figure 3

Figure 3



The patient was posted for EUA(Fig.4). Haematocolpos was drained under GA and vaginal septum was resected. Two cervices were visualized with difficulty after the resection of vaginal septum(Fig.5). Patient was asymptomatic at 6 weeks follow up.

Figure 4

Figure 4



Figure 5





DISCUSSION

Mullerian duct anomalies are congenital anatomic abnormalities that arise from non development or non fusion or failed resorption of Mullerian ducts. They are particularly important because they are associated with an increased risk of infertility, menstrual disorders, and obstetric complications. It is fundamental to have a basic knowledge of the embryology of the female genital tract to understand this group of congenital anomalies. The first stage of mullerian duct development begins at an approximately 6 weeks gestational age when the paired mullerian ducts invaginate and then grow caudally and cross over the Wolffian ducts to meet at the midline. The subsequent three phases (fusion, resorption, and vaginal induction) proceed in an orderly fashion from the 9th to the 22nd gestational week. Mullerian duct development occurs in close association with the development of the urinary system, and this explains the frequent association of anomalies of these two systems.

The Uterovaginal anomalies are classified by American Fertility Society in which class 1 is dysgenesis of Mullerian ducts, class 2 is disorders of vertical fusion of Mullerian ducts, class 3 is disorders of lateral fusion of Mullerian ducts which can be symmetric or asymmetric, and class 4 is unusual configurations of vertical-lateral fusion defects. Our case belongs to class 3 asymmetric variety (Fig.6)

Figure 6

Figure 6



Uterus didelphys is most often recognized as a part of a syndrome associated with obstructed hemivagina and ipsilateral renal agenesis. There is often delay in diagnosis when compared to complete obstruction (imperforate hymen, transverse vaginal septum) due to the fact that patients with obstructed hemivagina have periods ⁶. MRI is the most reliable non invasive modality for evaluating Mullerian anomalies⁷. It provides accurate preoperative evaluation and also useful in detecting coexisting urinary anomalies like renal agenesis, hypoplasia, dysplasia, ectopic ureters. Thus early and accurate diagnosis of this syndrome is important so that adequate and prompt surgical (excision

of the septum) can provide adequate relief and prevent complications like endometriosis. Successful obstetric outcome with this syndrome is 60%.

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